Annals of Physical and Rehabilitation Medicine 58 (2015) 183–185

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Elsevier Masson France EM|consulte www.em-consulte.com

Letter to the editor

# Motor and neurocognitive recovery in the syndrome of the trephined: A case report

ARTICLE INFO

*Keywords:*
Syndrome of the trephined
Sinking skin flap
Craniectomy
Cranioplasty

## 1. Introduction

Approximately, 1.4 million traumatic brain injuries occur each year in the United States. Among these incidents, 1.1 million result in emergency department visits, 235,000 result in hospitalizations and 50,000 result in death. This does not include an additional estimated 200,000 who are treated in outpatient settings, nor those treated at military facilities both domestic and abroad [1]. Decompressive craniectomy (DC) is frequently used for the treatment of medically refractory intracranial hypertension for patients with severe traumatic brain injury (TBI) [2]. Additionally, DC has been found to reduce mortality rates and improve outcomes in patients with malignant middle cerebral artery infarction, which accounts for 10% of stroke patients [3].

The syndrome of the trephined (ST), also known as the "sinking skin flap syndrome", is a disorder of delayed neurological deterioration [4]. By convention, ST refers to the development of those symptoms that are reversible after cranioplasty [5]. This is a complication that occurs in patients with large cranial defects following a DC. Related clinical deterioration occurs weeks to months after the surgery. The symptomatology and clinical signs can include: focal neurological deficits, aphasia, headache, dizziness, lethargy, irritability, inattention, memory problems, depression, anxiety, mood swings, behavioral disturbances, seizures, mutism, worsening hemiplegia, hemi-neglect, and diabetes insipidus [4–7] As these symptoms are non-specific, early diagnosis is difficult to establish, which can lead to significant neurological decline.

## 2. Case description

A 21-year-old male bicyclist was brought to the emergency department after being struck by a car. Upon presentation, the patient had a Glasgow Coma Scale (GCS) score of 11, a left hemotympanum, a fixed and dilated left pupil, and flaccid paralysis of the right upper extremity (RUE). The patient was sedated and intubated for airway protection and agitation management. Initial non-contrast head computed tomography (NCHCT) revealed a left epidural and subdural hematoma, diffuse subarachnoid and punctate intra-parenchymal hemorrhages, and a left temporal bone fracture. The patient required an emergent, left-sided DC to relieve his high intracranial pressure (ICP). After medical stabilization in the intensive care unit (ICU) and, subsequently, in an acute medical/surgical ward, the patient was transferred to an acute TBI rehabilitation unit.

His initial exam on the rehabilitation service revealed a large left craniectomy defect with a sunken skin flap. Motor testing revealed 3/5 strength in the left upper extremity (LUE) and flaccid paralysis in the RUE with spasticity noted at the elbow. He had poor sitting and standing balance. He had poor attention and inconsistently followed one-step commands with findings of motor apraxia, fluent aphasia, severe dysarthria, right unilateral body and spatial neglect, and asomatognosia. He required total assistance for all functional mobility and activities of daily living (ADLs).

Initially, he made modest but significant functional gains both cognitively and physically. He became able to ambulate on the unit with minimal assistance and perform ADLs with moderate to maximum assistance. He began to verbally communicate and interact with his environment and followed simple commands. He had minimal improvement in RUE strength and tone continued to fluctuate. About three weeks into his admission, he reached a functional plateau. Over the next month, he functionally declined cognitively and physically, developing mutism, requiring greater assistance with ADLs, and returned to use of a wheelchair for mobility. Follow-up imaging and medical work-up revealed no clear findings to explain his decline or lack of progression, but did reveal a sunken craniectomy defect and associated shift in brain parenchyma. After review and discussion with neurosurgery and the patient's family, the decision was made to proceed with a cranioplasty to improve his clinical status and functional recovery. Non-contrast head CT scans before and after the cranioplasty procedure are shown in Fig. 1.

Following the cranioplasty and medical stabilization, the patient was re-admitted to the rehabilitation service. The patient demonstrated immediate improvement. He had improved functional mobility, tone, strength, attention, awareness, memory, speech, social interaction, and emotional range. He began to communicate verbally and follow commands. He ambulated 1000 feet and negotiated 20 steps with minimal assistance to supervision. His AROM in both upper extremities were within functional limits. He was able to perform transfers at a contact guard to supervision level, ambulate with supervision, and perform activities of daily living at a supervision to moderate

http://dx.doi.org/10.1016/j.rehab.2014.11.003

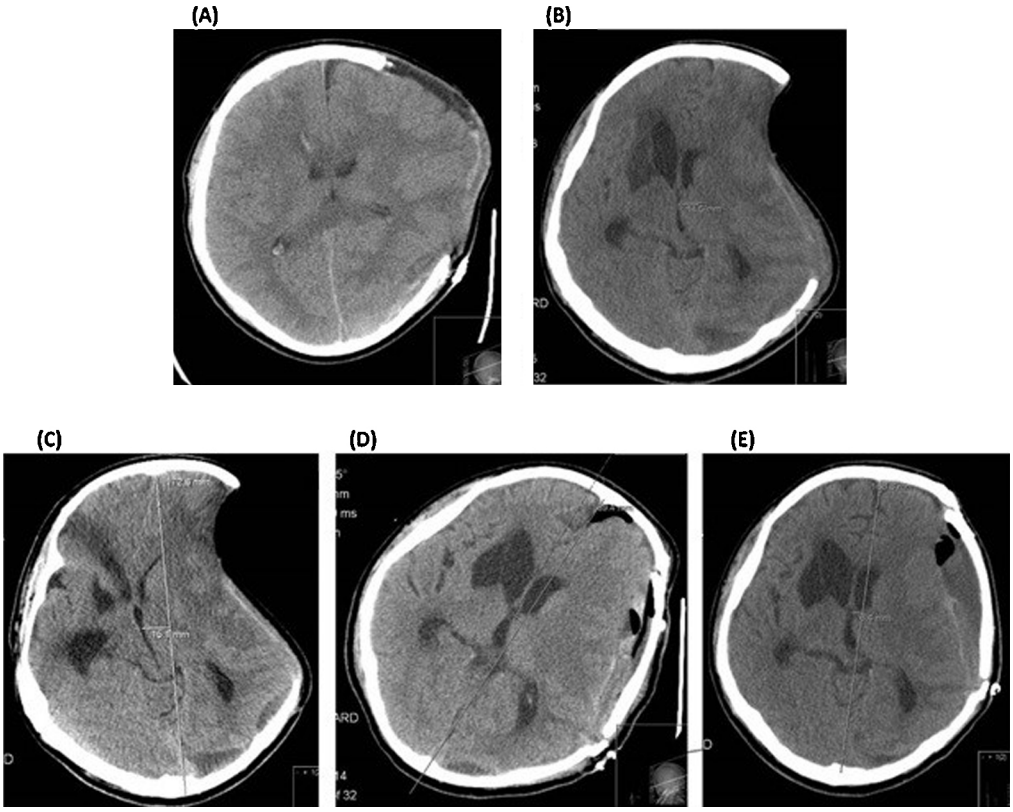


**Fig. 1.** Chronological NCHCT images of the clinical course: (A) 10 days post-craniectomy; (B) 38 days post-craniectomy (admission to rehabilitation unit, revealing a midline shift and sunken skin flap); (C) 64 days post-craniectomy (plateau/decline, revealing slight worsening of midline shift); (D) 3 days post-cranioplasty (demonstrating reversal of the sunken skin flap and resolution of shift); (E) 14 days post-cranioplasty (post-surgical follow-up, revealing stability).

assist level. After a 3-month course of rehabilitation, he was discharged home with his family.

## 3. Discussion

The most common cause of death and disability from TBI is high intracranial pressure (ICP) [8], resulting from intracranial volume expansion against a fixed, bony cranial vault. This may lead to herniation through the falx cerebri, tentorium cerebelli, and foramen magnum, with the potential to result in death [8]. It may be due to mass lesions, edema, or hemorrhage. Initial treatment focuses on maintaining normothermia, euvolemia, and moderate hypocapnia. Other interventions include a reverse Trendelenburg position, therapeutic cerebrospinal fluid (CSF) spinal tap, and administering of pharmacotherapy such as mannitol, barbiturates, and other blood pressure-modulating agents [9].

A DC allows for intracranial volume expansion without herniation risk by removing a portion of the skull [8]. The skull flap is not replaced immediately to allow time for the intracranial volume to decrease, thus effectively decreasing ICP. A DC typically requires subsequent admission to an ICU for neurosurgical monitoring, followed by an acute ward stay for medical stabilization. Rehabilitation is also typically required as a necessary and intermediate step for discharge, mitigating the impairments secondary to the underlying disease process, post-surgical deconditioning and functional decline secondary to immobility. In clinical practice, timing recommendations for DC reversal and reconstructive cranioplasties are highly variable. Some studies insist that DC should no longer be considered as an appropriate second-tier therapy for elevated ICP, because non-surgical treatment have been found to be as effective for improvement of outcomes, and because a decompressive craniectomy procedure is very invasive and is associated with substantial medical complications in as many as 50% of patients [2]. Complications from the procedure aside from ST include herniation through the craniectomy defect and injury to the herniated cortex, subdural effusion, hydrocephalus, and post-traumatic seizures [10].

The cause of ST is controversial but has been attributed to a number of factors. A known cause is local in-folding of the scalp or scarring at the craniectomy site between the overlying skin and dura, which exerts direct pressure on the brain [11] Atmospheric pressure also exerts a force against the concave sinking skin flap and underlying brain tissue, influencing CSF flow. A previously documented study posited that derangements in CSF flow cause leakage and transgression of CSF and resultant edema into contused parenchyma, which is analogous to the transependymal CSF flow and edema in patients with normal pressure hydrocephalus. The study found a high incidence of CSF flow derangements in the form of CSF hygromas in patients with a delayed motor deficit occurring after DC, a subset of ST known as "motor trephine syndrome" [6]. Additionally, cerebral blood flow (CBF) may be altered by local temperature fluctuations due to the lack of insulation previously provided by the skull, compromising cerebral perfusion and venous drainage and resulting in ischemia.

There are few conservative and non-operative treatment options for presumed ST. Alterations in head posture, particularly while in the Trendelenburg position, provide restoration of the curvature of the scalp and is strongly related to improvement in neurological status. A previously documented case describes a patient with ST whose head was lowered by 10 degrees relative to the horizontal plane of the table, resulting in a GCS score increase from 8/15 to 12/15 and a reversal of the sunken scalp flap over several hours. In another documented case report, an ST patient experienced prompt reversal of dysarthria and right hemiparesis after transition to a Trendelenberg position. From a surgical

management standpoint, aside from the aforementioned cranioplasty, another procedure is lysis of scar tissue with an operative goal to improve CBF [7].

Cranioplasty is considered the most effective and permanent treatment of ST [5]. Repair of the defect presumably removes the mechanical and atmospheric pressure exerted on the brain parenchyma and restores normal CBF and CSF hydrodynamics. Radiological studies including xenon CT, perfusion CT, and dynamic CT imaging have shown improvement in CBF following cranioplasty at the site of the DC [7]. Multimodal functional imaging reveals significant changes in functional networks and cerebral metabolism before and after cranioplasty [12]. It has been theorized that restoration of the intracranial environment may trigger a switch from the glycolytic pathway towards a more efficient means of producing ATP within neurons, resulting from a protein phosphorylation mechanism that enhances cerebral metabolism and optimizes neuronal plasticity [6,12]. Complications of cranioplasty do occur, however, and include sudden death following autologous cranioplasty, infection, bone flap resorption [10].

In summary, this case describes a patient suspected to have ST and who underwent a cranioplasty and benefited from a significant neurological recovery. Similarly, reported cases in the literature have involved patients with acute medical or functional decompensation [12]. From a clinical perspective, it is crucial to recognize the early signs of ST in order to quickly and efficiently refer to a neurosurgical team for cranial repair [6,12]. Early detection may prevent unnecessary treatments and avoid apportioning limited rehabilitation resources to patients who cannot derive benefit due to the ST [12]. This may lead to improved treatment algorithms post-DC, as no studies to the authors' knowledge have been conducted on primary and secondary prevention of ST.

Furthermore, recent advances in cranioplasty implant fabrication have facilitated operative treatment and demonstrated improved outcomes. There are now customized cranioplasty implants that utilize 3-dimensional printers and polymethylmethacrylate casting. These prefabricated implants reduce surgical time, decrease the potential complications, assure optimum aesthetic results, and may have advantages with respect to cost, manufacturing time, and complexity of the procedure [13].

## 4. Conclusion

Physiatrists caring for TBI patients after a DC should be aware of the clinical features of ST and recognize the significance of a functional plateau and/or decline. With this understanding, physiatrics can recommend earlier intervention for cranioplasty in order to avoid further decline attributed to ST. With the trend toward earlier transition of care from acute medical to acute rehabilitation, the rehabilitation inpatient unit is where the delayed onset of ST will increasingly be seen. Assuming resolution of intracranial edema and hemorrhage, and neurosurgical stability, early cranioplasty should be considered in the rehabilitation setting in craniectomy patients with acute, clinical decompensation. Care must be taken as the earliest signs may come in the way of functional plateau and/or subtle decline. It behooves the physiatrist to consider a neurosurgical consult to discuss early cranioplasty as a viable management option. Further research and detailed management algorithms are needed to optimize care.

## Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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Received 18 September 2014
Accepted 26 November 2014